

Case Report

Non-Hodgkin's lymphoma as a cause of intussusception: a case report

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ABSTRACT

Intussusception of the bowel is defined as the telescoping of a proximal segment of the gastrointestinal tract within the lumen of the adjacent segment. This condition is frequent in children and presents with the classic triad of cramping abdominal pain, bloody diarrhea and a palpable tender mass. Secondary intussusception is caused by organic lesions, such as Meckel's diverticulum, benign and malignant lesions, metastatic neoplasms or sometimes iatrogenically. Surgery is the definitive treatment of secondary intussusception. Formal bowel resection with oncological principles is followed for every case where a malignancy is suspected. A ten-year-old female presented to the casualty with severe pain in abdomen associated with 1 episode of blood in stools diagnosed with intussusception. She was taken for emergency laparotomy, and no lead point was identified. She presented with recurrent intussusception and was taken for re-exploration, when a definitive approach was opted for, and she underwent a resection of the obstructed bowel at the ileo-caecal junction and an ileo-ascending anastomosis. On histopathology, the ileo-caecal junction showed a proliferative, circumferential mass which was confirmed to be a diffuse large B cell lymphoma. It is uncommon for pediatric intussusception to result from a neoplasm. It takes imaging modalities to make the diagnosis. Surgical intervention is required as soon as a suspicion is raised in order to avoid consequences such as gut necrosis and ischemia.

Keywords: Intussusception, Pediatric-surgery, Diffuse-large-B-cell-lymphoma, Ileo-caecal-junction, Bowel-resection, Neoplasm

INTRODUCTION

Intussusception is defined as the telescoping of a segment of the gastrointestinal tract into an adjacent one. A demonstrable etiology is found in 70% to 90% of cases in adult patients, and about 40% of them are caused by a primary or secondary malignant tumor.

Intussusception occurs when a more proximal portion of bowel (intussusceptum) invaginates into the distal bowel (intussusciens). The pathomechanism is thought to involve altered bowel peristalsis at the intraluminal lesion, which is then a lead point for the intussusceptum.¹

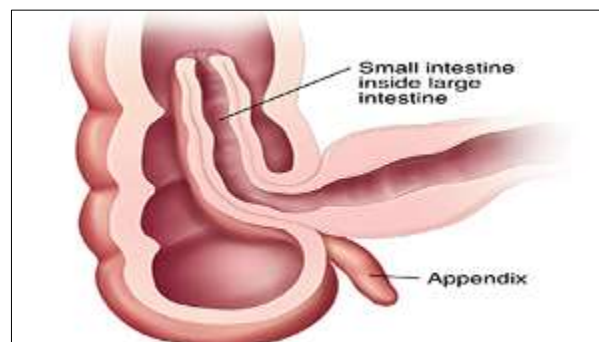


Figure 1: Illustrated image of intussusception.²

Commonly seen in children, adult intussusception represents only 5% of all the cases of intussusception, according to Azar et al.¹ The pathophysiology is thought to result from an imbalance in the longitudinal forces along the intestinal wall, which can be brought on by a disorderly pattern of peristalsis (such as an ileus during the postoperative period) or a mass functioning as a leading point. This causes the mesenteric arteries to compress and angulate, which lowers perfusion, venous congestion, and intestinal wall edema, potentially leading to ischemia and intestinal necrosis.³ Benign lesions make up the bulk of lead points in the small intestine. Up to 30% of small intestine intussusception cases are caused by malignant tumors.³ Large bowel intussusception accounts for 63% to 68% of cases and is more likely to have a malignant cause.⁴

With 5% to 20% of all cases, the gastrointestinal system is the most often impacted extranodal location by lymphoma. However, primary gastrointestinal lymphoma is extremely uncommon, making for about 1% to 4% of all gastrointestinal cancers.⁴

Histopathologically, almost 90% of primary gastrointestinal lymphomas are B-cell non-Hodgkin's lymphoma (NHL), followed by T-cell NHL and Hodgkin's lymphoma (HL).⁴

Gastrointestinal lymphoma is usually secondary to widespread nodal diseases. The most frequently affected sites are the stomach (46.47%), followed by the small bowel (25%) and colon (8%).⁵

We saw one such case of extra nodal Hodgkin lymphoma causing an intussusception in a 10-year-old.

CASE REPORT

Ten-year-old girl presented to the casualty with chief complaints of severe pain in abdomen associated with 1 episode of blood in stools. There was associated fever which was relieved on medication. There was a previous history of intermittent episodes of colicky pain which resolved spontaneously. The patient was brought in attack of severe colic.

An ultrasonography, followed by a contrast enhanced computed tomography (CECT) scan (A+P) showed large ileo-colic and colo-colic intussusception, the caecum and terminal ileum along with ascending colon (intussusceptum) is seen entering into the transverse colon (intussusceptiens). Multiple lymphnodes were identified along the ileocaecal vessels. Edema was noted in wall of the intussusceptum.

Patient was taken for emergency exploration on 16 March 2024. The intussusception was reduced till serosal continuity was observed, under the guidance of the pediatric surgeon in our department, Dr Anjali Chitale (Professor, Dept of General Surgery).

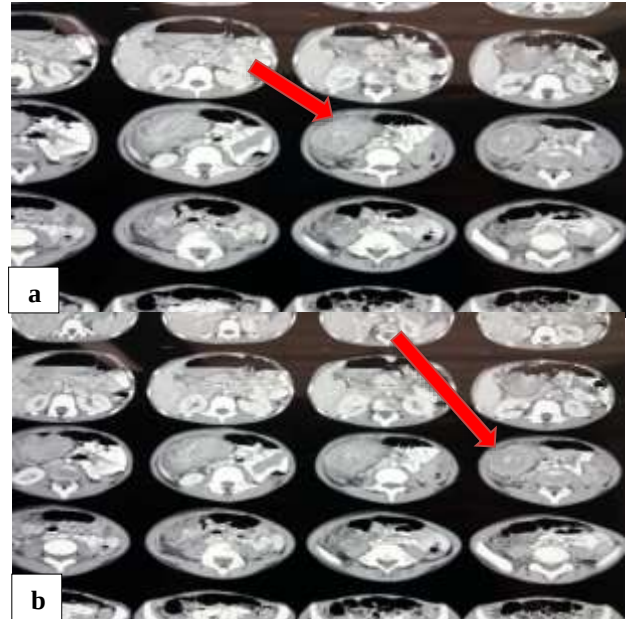


Figure 2 (a and b): CT films showing bowel with bowel appearance suggesting intussusception shown in red arrows.



Figure 3: Intraoperative photo showing serosal continuity of ileocaecal junction.

Patient recovered from surgery, and was discharged after passing stools. A repeat ultrasonography scan was done, which was suggestive only of caecum size 4.5 cm, and minimal ascites with cholelithiasis.

Patient presented again with complaints of colicky pain in abdomen after 17 days, and underwent another ultrasonography for the same, which was suggestive of a recurrent intussusception. This was confirmed on CECT scan (A+P).

When the patient was taken for re-exploration on 11 April 2024, a palpable circumferential mass at the ileo-caecal junction was discovered during the laparotomy. As a result, the decision to conduct an ileo-caecal junction resection with an ileo-ascending anastomosis (with a 5 cm margin on both sides) was taken. The resected specimen was sent for histopathology.

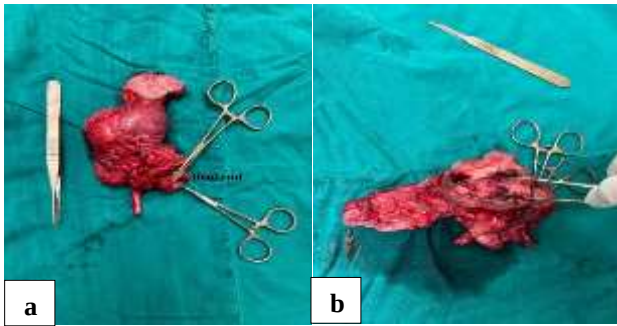


Figure 4 (a and b): Specimen photos.

The histopathology report was suggestive of lymphoma, which was confirmed to be a diffuse large B cell lymphoma on immunohistochemistry markers. The slides are seen below with different magnification.

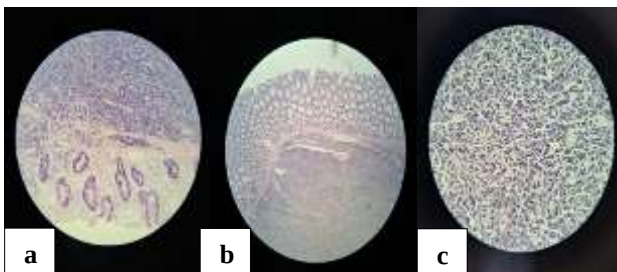


Figure 5: Histopathology slides as seen above, (a) low power view showing small to medium size atypical lymphoid cells infiltrating the mucosa; (b and c) high power view showing monomorphic population to atypical lymphoid cells.

In the subsequent follow up months, she is receiving timely treatment for the diffuse large B cell lymphoma, and at present has no complaints.

DISCUSSION

In three fourth cases, the cause of intussusception in children is not known. Pathological lead points have been identified in 4% of cases, most common being Meckel's diverticulum, lymph nodes and intestinal polyps. Rarely, lymphomas, duplication cysts, parasites, hematomas, vascular malformation, sutures and staples or any intraluminal foreign body are reported.

Two thirds of the non-Hodgkin's lymphomas present with lymphadenopathy. The remaining 1/3rd present with symptoms relating to the involvement of extra nodal sites (e.g. gastrointestinal tract, Waldeyer's ring, brain, skin,liver, testes, and kidney).⁶

The gastrointestinal tract is the most common extra nodal site affected by lymphoma, accounting for 5% to 20% of all cases. Primary gastrointestinal lymphoma, however, is very rare, constituting only about 1% to 4% of all gastrointestinal malignancies. Gastrointestinal lymphoma is usually secondary to widespread nodal diseases. The

most frequently affected sites are the stomach (46.47%), followed by the small bowel (25%) and colon (8%).^{5,6} Histopathologically, almost 90% of primary gastrointestinal lymphomas are B-cell non-Hodgkin's lymphoma (NHL), followed by T-cell NHL and Hodgkin's lymphoma (HL).

According to most reports, surgery alone can be curative if the tumor is resectable.¹¹ Chemotherapy is clearly indicated in all patients with disseminated disease. Multi-agent chemotherapeutic strategy is warranted for advanced stage intestinal lymphoma with multifocal presentation of MALT lymphoma. Chemotherapy regimen, consisting of Rituximab alone or purine nucleoside analogs with Rituximab, can be applied to those ineligible for stem cell transplantation.⁶ Radiation therapy in addition to surgery has been thought to be unnecessary if the tumor has been completely resected.⁵

Management of recurrent intussusception typically involves non-operative reduction initially, with surgical intervention reserved for specific circumstances. Recurrence rates vary, with studies reporting rates around 8% to 13.8%.⁸ Non-operative methods like hydrostatic or pneumatic enema show high success rates in initial and recurrent episodes. For recurrent cases, success rates for non-operative reduction remain high, ranging from 92% to 96.2%.⁹

Surgical intervention becomes necessary if non-operative methods fail, or if there's suspicion of a pathologic lead point (PLP), found in about 9.3% of recurrent cases.⁹ PLPs include colonic polyps and Meckel diverticulum. The overall approach to recurrent intussusception mirrors that of primary cases, emphasizing conservative management initially and surgical options for failures or suspected PLPs. Despite recurrences, mortality remains low, and outcomes are generally favorable with appropriate management tailored to each recurrence episode as if it were a primary occurrence.

In general, the majority of lead points in the small intestine consist of benign lesions. Rarely, there may be benign lymphoid hyperplasia of the ileo-caecal junction causing ileo-caecal-colic intussusception.¹⁰ Malignant lesions account for up to 30% of cases of intussusception in the small intestine. Intussusception occurring in the large bowel is more likely to have a malignant etiology and represents 63% to 68% of cases.^{10,11} It is always better to keep a malignant pathology in mind while operating a palpable lump or growth.

CONCLUSION

Surgical management of intussusception during exploratory laparotomy often requires imaging for accurate diagnosis, particularly when the lead site is not palpable externally. Considering lymphoma and other neoplasms as potential causes, underscores the importance of prompt surgical intervention to prevent complications

like gut necrosis and ischemia. Adjuvant chemotherapy is crucial for gastrointestinal NHL treatment alongside surgery to eliminate the tumor. Emphasizing caution in treatment and regular postoperative follow-up can reduce the incidence of advanced, disseminated disease and lower associated mortality rates. This study advances understanding by highlighting the necessity of a precise clinical approach followed by combining surgery and chemotherapy, thereby improving outcomes through early intervention and vigilant monitoring.

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